

CHILD NEUROLOGY (GENERAL PEDIATRIC NEUROLOGY)

by measuring nasal nitric oxide (nNO), a screening test for Primary Ciliary Dyskinesia (PCD). *Study design:* We measured nNO levels of 26 children with congenital midline CNS defects. We evaluated the effect of age, gender, and anomaly (brain, spinal cord, or combined) on measurements. We compared our results to the previously established normal range (153.6-509.9 nL/min), and to the cutoff for PCD (77 nL/min). *Results:* The range for nNO in our cohort was 56.5-334.7 nL/min, with age, gender, and anomaly not having a significant effect. The overall mean, 217.7 nL/min, was significantly lower than that of normal children, 314.51 nL/min ($p < 0.01$). Four subjects (15.4%) had nNO levels below the lower end of normal, with two (7.7%) having values fitting the cutoff for PCD. *Conclusions:* We report an association between ciliary dysfunction and isolated midline neuroanatomical defects, not in context of any known syndrome. This suggests that genes causing isolated CNS defects, may be implicated in the function of cilia. Longitudinal studies are required to investigate whether children with abnormal measurements suffer from any respiratory sequelae.

P.005

Utilization of transition care management plans to facilitate transition of adolescents with epilepsy into the adult healthcare system

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Background: For adolescents with epilepsy, there is often a poor system in place to meet their individualized transition needs. Our objectives were 1) to develop epilepsy-specific transition care management plans (TCMPs) to ensure access, and attachment to adult healthcare providers, and 2) to identify strategies for providing support during the transition period, including through the development of physician and patient (or caregiver) navigated web-based tools, resources and recommendations for health system improvements. *Methods:* Physicians and nurses with expertise in areas including adult and pediatric epilepsy, family medicine, psychiatry, and varied allied health professionals were engaged to generate epilepsy-related TCMPs. *Results:* Through an iterative process spanning the course of over a year, TCMPs were developed to cover areas including: treatment responsive and resistant epilepsy, ketogenic diet, epilepsy surgery, women's issues, mental health, and psychosocial aspects of epilepsy. The TCMPs referenced established guidelines and best practices in the literature wherever possible. Caregiver roles and responsibilities were outlined, remaining cognoscent of available provincial resources. *Conclusions:* Epilepsy specific TCMPs can be developed through a collaborative approach between pediatric and adult healthcare providers, easing the patient experience, creating educated accountability, and providing a forum to identify and address gaps of care in adolescents with epilepsy.

P.006

Increased healthcare services utilization in the tuberous sclerosis complex population in Quebec

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Background: Tuberous sclerosis complex (TSC) is a neurocutaneous syndrome that can present with many disabling neurological symptoms, the most common being seizures. Although it is a chronic systemic syndrome, healthcare utilization and long-term outcome of subjects with TSC are not well defined. The goal of this study was to evaluate the direct cost and long-term outcome of TSC compared to other forms of epilepsy and healthy controls. *Methods:* Our provincial health care database was interrogated to determine use of medical services by patients with TSC, epilepsy and healthy controls from 1996-2011. Data on demographics, outcomes and health care utilization were analyzed. *Results:* 1004 TSC, 41,934 with epilepsy and 41,934 controls were identified. The prevalence of TSC was 1/7,872 compared to 1/189 for epilepsy. TSC experienced more hospitalizations, medical visits and prescription drug use, resulting in higher total health care costs. Their most common admission diagnosis was seizures and age at death was significantly lower: 61.3 years old for TSC vs 69.6 and 76.6 years old for epilepsy and controls, ($p < 0.001$). *Conclusions:* TSC subjects have a significantly higher burden of disease than other subjects with epilepsy. These results stress the need for specialized services in this population through the lifespan.

P.007

Topographical orientation as a model of plasticity in children with perinatal stroke

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Background: Children with perinatal stroke go on to develop most cognitive skills (e.g. language) due to brain plasticity; however, their performance is usually poor when compared to age-matched controls, indicating a reduced potential compared to uninjured children. To date, how plasticity after early injury affects the development of complex cognitive skills remains uncertain. Here, we use topographical orientation, which relies on integration of several cognitive processes underlain by widespread neural networks, as a model to test plasticity in complex behaviour. *Methods:* Children with perinatal stroke and age-matched controls were tested with a neuropsychological battery and a novel navigation task. In addition, for each patient, we obtained the most recent MRI scan to assess the effects of lesion characteristics on performance at the navigational task. *Results:* Children with history of injury performed worse than controls, and their scores were not different based on lesion's laterality, location or functional region affected. In particular, involvement of regions known to contribute to spatial orientation did not result in significantly decreased performance. *Conclusions:* As seen in other